Capgras' syndrome with organic disorders

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Summary: Capgras' syndrome, one form of the delusional misidentification syndromes, is described. Three patients with the syndrome are reported. The first had a right cerebral infarction, the second had nephrotic syndrome secondary to severe pre-eclampsia in the puerperium, and the third had uncontrolled diabetes mellitus with dementia. Evidence is reviewed regarding an organic aetiology for Capgras' syndrome. We conclude that, when the syndrome is present, a thorough search for organic disorder should be made.

Introduction

Capgras' syndrome was described by Capgras and Reboul-Lachaux in 1923,1 and has exercised a fascinating hold over medical writers ever since, if the number of published reports is anything to go by.²⁻⁴ Capgras, whose name was attached to the syndrome by Levy-Valensi, sused the term 'illusion des sosies' to describe the phenomenon whereby a patient believed that someone, usually a loved one, had been replaced by an identical looking imposter. 'Sosie' is the French word meaning double, and derives from the Greek Sosias who was servant to Amphitryon. According to legend, Zeus, everlustful, desired to have sexual intercourse with Alcmene, who was inconveniently devoted to her husband Amphitryon. While the latter was at work, Zeus assumed an identical form to him, and tricked Alcmene into making love with him, as a result of which Hercules was conceived.

The real Amphitryon returned in the evening, and, surprising Alcmene with apparently renewed ardour, proceeded to conceive Hercules' twin brother, Iphicles, who, one suspects, spent his life failing to live up to his brother's exploits, since history says little more about him. The servant Sosias, who might otherwise have given the game away, was removed by Zeus and replaced by Mercury, who assumed Sosias' form. Mercury could be relied upon to let Zeus get away with his deception.

We report three cases of this interesting syndrome in which organic factors most probably were aetiologically significant.

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Case reports

Case 1

A 69 year old widow barricaded herself into her flat one week after returning from holiday. She had become uncharacteristically irritable and suspicious.

The day before, she had set a small fire, but refused to allow the firemen in, claiming they were not real firemen. On the day of assessment, she had poured a bucket of water over a group of elderly ladies who were passing under her bedroom window. She said that these ladies, who were, in reality, known to her, were not her real neighbours.

When her mental state was finally examined (her door needing to be broken down) the most striking finding was her conviction that her sister had been replaced by an impostor who looked the same, but who gave away, by subtle differences of gesture and mannerism, the substitution. The impostor was organizing the persecution, which took the form of objects in her flat being inexplicably moved, a laser beam being directed at her (this being her explanation for a shaft of sunlight shining through her window), and the invasion of her privacy which included the breaking down of her front door.

In all other respects her mental state was normal. Detailed cognitive examination, including memory and orientation, was normal. Physical examination revealed a sprightly, normotensive lady with an old tuberculous arthrodesis of her left knee, and signs of a deep vein thrombosis affecting the left leg. There were no other abnormalities, and, in particular, neurological examination was entirely normal. She was right-handed.

After admission to hospital she was treated with neuroleptic medication, initially droperidol, then trifluoperazine. After 10 days her symptoms began to disappear, and by 3 weeks she was her normal self again. She was perplexed at how she could have believed what she now accepted to be absurd.

Routine physical investigations were all normal. Erythrocyte sedimentation rate was 42 mm in the first hour. A venogram demonstrated thrombosis of the left internal iliac vein. A computed tomographic (CT) brain scan showed a small infarct in the right internal capsule. Appearances were consistent with the infarct having occurred in the preceding 3 weeks. As a result of the investigations, she was started on heparin, and subsequently warfarin. Her subsequent progress has been unremarkable, apart from predictable neuroleptic side effects. All medication was discontinued after 2 months and she has remained well, physically and mentally, in outpatient follow-up over 9 months.

Case 2

A 31 year old Philippino woman underwent elective Caesarean section at 30 weeks' gestation because of pre-eclampsia causing severe hypertension and poor fetal growth. This was her first pregnancy and she had previously been entirely well medically and psychiatrically.

Immediately after delivery, her mental state was normal. Physical examination revealed peripheral oedema and hypertension. She had proteinuria and low serum albumin.

She remained on the post-natal ward, recovering from her Caesarean section, apparently uneventfully. Nine days after the operation she suddenly became low in mood, 'confused and strange' on the telephone to her husband, and could not find her way round the ward. She rapidly became agitated, shouting, screaming and hitting out, such that parenteral sedation was required.

She was then transferred to the local psychiatric hospital. On mental state examination, she was calm, co-operative and without affective disturbance. The striking abnormality was the presence of the Capgras' delusion concerning her husband. She said he had been replaced by an impostor. She examined her husband's face and pointed out very subtle differences, such as an extra furrow on his brow, which made her realise that he was not really her husband, but a virtually identical double. Her cognitive examination revealed disorientation in place and impaired short term memory. The only physical abnormalities were peripheral oedema, proteinuria and hypertension, with blood pressure 190/120 mmHg. Fundoscopy remained normal. Her hypertension persisted despite treatment with labetalol and nifedipine. She was started on droperidol. The following day she had a probable grand mal convulsion. Her Capgras' delusion persisted for 3 more days and then disappeared,

with entirely normal mental and cognitive state thenceforth.

Detailed investigation confirmed nephrotic syndrome. Renal biopsy was consistent with IgA nephropathy. Computed tomographic brain scan was normal.

Over the next several weeks there was rapid improvement in her medical condition, the nephrotic syndrome resolving and the hypertension disappearing. Eight weeks after delivery of her baby, she was entirely well mentally and physically, off all medication, and with normal blood and urine protein levels.

Case 3

A 74 year old woman with long standing diabetes mellitus, but otherwise well, was referred for assessment of deteriorating memory and episodes where she failed to recognize her husband. Her diabetes, under insulin control, had been brittle, and there were repeated hypoglycaemic episodes in the 2 years before assessment.

Fifteen months before assessment, her first episode of misidentification of her husband occurred. She suddenly asked what he was doing in the house, when her real husband was out in the garden. She knew that the person in the room looked like her husband, but, equally knew that he was not. This episode lasted several hours, was not associated with any other obvious symptoms, and she was amnesic for it afterwards. Some months later, a second, similar episode occurred, and episodes then occurred with increasing frequency, until they were happening on most days. Each episode lasted from an hour up to 36 hours.

There had also been gradual deterioration in memory over some months. She frequently misplaced things and put them in strange places. She occasionally left taps running, and cookers burning. She could not reliably remember messages or shopping lists.

Physical examination was normal, apart from signs of mild neuropathy and diabetic retinopathy. The only abnormalities of mental state were mild impairment of short term memory, abstract thinking and judgement. When first seen, she correctly identified her husband.

Routine investigations were normal. Random blood glucose estimations were consistently elevated. Electroencephalogram (EEG) was consistent with ischaemic disturbance. CT brain scan showed widening of the lateral and third ventricles, consistent with mild atrophy. There was evidence of microvascular change. Formal psychometric testing confirmed the memory impairment, and suggested an organic aetiology. While under investigation, the patient demonstrated the sudden onset of the Capgras' delusion. In the middle of the

afternoon she told her husband, with whom she had previously been talking normally, that he was not her real husband, but was adamant that she had never seen him before. This belief persisted for several hours. She was otherwise unchanged in her examination. A blood sugar taken during this episode was 17 mmol/l. During subsequent episodes, her husband performed BM-Stix estimations of blood sugar, and these were invariably raised.

After investigations were completed, the patient and her husband moved, and have been followed up by their general practitioner. She has developed worsening syptoms of dementia, and requires 24 hour nursing care, 3 years after the onset of her Capgras' syndrome.

Discussion

In all three cases, there is an association between an organic disorder and Capgras' syndrome. The first patient suffered a small right cerebral infarction which most probably coincided with the onset of psychiatric symptoms. An underlying pathology, such as neoplasm, in association with a deep vein thrombosis was not discovered.

In the second case, the patient had toxaemia, with worsening hypertension and nephrotic syndrome on the post-natal ward. The behavioural disturbance may have prejudiced her assessment, such that physical factors were not sufficiently explored. The sustained severe hypertension may well have produced alterations in cerebral perfusion, even if permanent neuronal damage did not occur, as suggested by the normal CT scan.

The third patient, interestingly, demonstrated an intermittent Capgras' syndrome, of sudden onset and abrupt cessation, on a background of poorly controlled diabetes and early dementia. Hyperglycaemia was demonstrated during the episodes. We surmise that a limited functional cerebral reserve was further impaired by the metabolic consequences of hyperglycaemia, although other explanations, for example epileptic phenomena, are possible. It is worth noting that the diagnosis of Capgras' syndrome was not at first made in this patient. Instead prosopagnosia was diagnosed. Because she could, in fact, recognize faces, but merely had a delusion, or false belief, concerning the identification of her husband's face, correctly perceived, this diagnosis was incorrect. This distinction between prosopagnosia and Capgras' syndrome has been discussed.6

Any general consideration of the aetiology of Capgras' syndrome must take account of allied misidentification syndromes, of which there are many. In the Fregoli syndrome, 7.8 honouring an Italian actor and mimic, an imagined persecutor is

seen to take the form of many different people, otherwise familiar. In the syndrome of subjective doubles, the patient believes there to be doubles of his own self exclusively, while in the syndrome of intermetamorphosis, the patient believes others have changed their shape or appearance.

The belief that a physical location has been duplicated, reduplicated paramnesia, was first described by Pick. ¹¹ Interestingly this has always been considered a 'neurological' syndrome, ¹² whereas the others are 'psychiatric'. The only significant difference is that, in the former, place is misidentified and duplicated, while in the latter, people are misidentified but not necessarily duplicated. In fact, Pick's two cases demonstrate both misidentifications, lending support to the idea that the two syndromes are intimately connected. Other conditions in which organic aetiology is more obvious include 'deja-vu', 'jamais-vu', and 'out of body' experiences, associated with temporal lobe disturbance, and autoscopy. ⁴

The 'double' clearly is a powerful concept. Many different cultures have myths and beliefs involving doubles. We have already mentioned the Greeks. Zeus' transformations alone rivalling the number of case reports of Capgras' syndrome in the modern literature. We could also refer to the ancient Egyptians with their concept of Ka, and Ovid's Metamorphoses in Latin culture. Less well known are myths from Irish, Indian, Indonesian and Australian cultures. 13 Literature, too, abounds with the theme of substitution and misidentification. Perhaps the clearest literary account of the Capgras' syndrome is by Dostoyevsky in 'The Possessed', describing the non-recognition by Marya Timofeyevna of her husband Stavrogin. She says 'you are like him, very like him, perhaps you are a relation - only (my husband) is a bright falcon and a prince, and you are an owl and a shopman'. 14 In 'The Double', Dostoyevsky describes Mr Golyadkin's battle with his own double. 15 Dostoyevsky himself suffered from temporal lobe epilepsy with psychiatric complications.

Not surprisingly, a number of psychodynamic theories have been put forward to explain Capgras' syndrome. 2,16 Increasingly, however, the evidence for an organic aetiology is becoming apparent. The degree of investigative vigour, and technological advances in brain imaging techniques influence the likelihood of demonstrating an organic disorder. In 1983, Berson² reported that 23% of cases identified in the English language literature had a clear organic disorder, the rest having schizophrenia or manic-depressive illness. In 1987, Signer⁴ reported that 40% had evidence of an organic disorder.

Joseph¹⁷ studied 29 patients with single or multiple misidentification syndromes, using one or more of electroencephalography (EEG), evoked responses (ER), computerized tomography (CT),

neuropsychological testing (NPSYCH), brain electrical activity mapping (BEAM), position emission tomography (PET), and single photon emission computed tomography (SPECT) of the brain. He found a very high percentage of abnormalities in at least one of CT, EEG, ER and BEAM. In general the findings demonstrated diffuse cortical dysfunction, although one patient had right hemisphere dysfunction alone, an abnormality previously suggested as being aetiologically significant in Capgras' syndrome.

Capgras' syndrome has been reported with various organic disorders, including non-specific right cerebral dysfunction^{18,19,20} subarachnoid haemorrhage,²¹ head injury,^{22,23} temporal lobe dysfunction,²⁴ pseudohypoparathyroidism,²⁵ myxoedema,²⁶ and many other conditions.²⁷ Of particular interest regarding our cases is the scarcity of reports of Capgras' syndrome with puerperal psy-

chosis²⁸ or epilepsy.²⁹

Joseph¹⁷ proposes that the abnormality in the misidentification syndromes involves disconnection between right and left hemisphere cortical areas that decode afferent sensory information pertaining to 'orientation', be it for time, place or person. His hypothesis, while speculative, is similar to the theory of Green³⁰ who explains auditory hallucinations in terms of a neuropsychological defect in the processing of language between right and left hemispheres.

In conclusion, the evidence favours the likelihood of an organic aetiology for many cases of Capgras' syndrome. We have presented three cases in which organic factors were clearly identifiable. We suggest that where any delusional misidentification is present, there should be a thorough search for organic pathology.

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